



pylorus or ileocecal valve and objects longer than 5 cm will not pass by the duodenal sweep.

Endoscopic removal is contraindicated if the object is located above the upper esophageal sphincter or if there is clinical or radiographic evidence of perforation. Objects located proximally to the upper esophageal sphincter should be removed by an otolaryngologist.³⁻⁵ Ingested sharp-pointed

objects have the highest rates of perforation, (up to 35%) and those within the esophagus, stomach or duodenum should be removed endoscopically on an urgent basis. Surgical intervention is considered if endoscopic removal fails, if the patient develops symptoms or if the object fails to progress over 72 hours. Laparotomy should be reserved for those who develop overt acute abdomen and in whom the object cannot be endoscopically retrieved.

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Congenital short pancreas

Introduction

Pancreatic anomalies are only rarely reported, but complete agenesis of the dorsal pancreas is extremely rare.¹ Partial agenesis mostly affects dorsal part, because it relies on different genes and signaling events from those of ventral pancreas.² Our case presented with recurrent episodes of pancreatitis. Investigations showed absent body and tail of the pancreas. Patient had no evidence of chronicity in the remaining pancreas.

Case report

A 27yr old male patient presented with history of abdominal pain in the epigastrium for the last two years. He had recurrent episodes of pain which used to increase with food intake. Twice he had severe pain which required admission to the hospital. He used to note that there is increase in pain after bouts of alcohol intake. The pain used to radiate to the back and last for hours. There was no history of trauma in the past. General examination was non contributory. Abdomen was soft and did not reveal any mass or free fluid.

His routine investigations were normal. Amylase and lipase levels were normal at presentation. His blood sugar and serum calcium were normal. His glucose tolerance test was normal. Triglyceride levels were normal. Ultrasound report showed bulky pancreatic head.. Even though he had significant history of alcohol intake, a CT scan was advised to rule out any structural abnormality. CT scan showed absence of body and tail of pancreas. MRI (**Figure 1**) and MRCP (**Figure 2**) showed complete absence of body and tail of pancreas (which develops

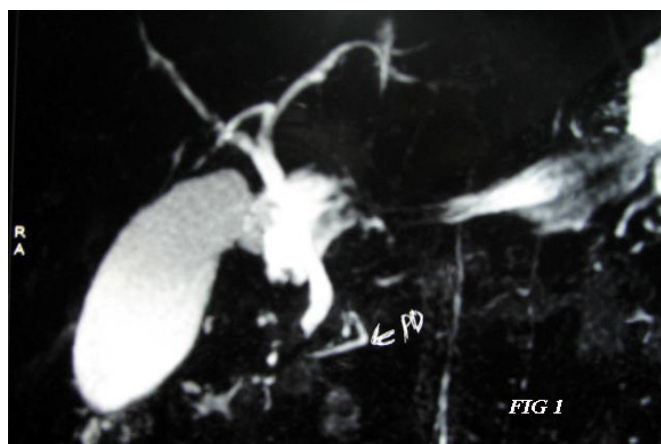


Figure 1: MRI showing small pancreatic duct



Figure 2: MRCP showing small pancreatic duct (ventral pancreas)

from dorsal pancreatic bud). There was no evidence of calcification or pancreatic duct dilatation.

He was advised to stop alcohol and was put on pancreatic enzymes. He is under follow up now. For the last six months he never had any abdominal pain. He is planned for sphincterotomy of pancreatic duct if he further develops recurrent pancreatitis, as there are occasional case reports of pain relief with pancreatic sphincterotomy.³

Aplasia, or varying degrees of hypoplasia, of the pancreas were classified histologically by Lumb et al⁴ into (1) complete (2) partial (a) involving both endocrine and exocrine tissue (b) involving endocrine tissue only (c) involving exocrine tissue only. According to the degree of immaturity of the dorsal pancreas development, hypoplasia of the pancreas is classified clinically into three types (1) total agenesis of the dorsal pancreas (2) hypogenesis of the body and tail (3) hypogenesis of the tail. In type 1, the major pancreatic duct can be identified on endoscopic retrograde cholangiopancreatography (ERCP) images, but not the minor pancreatic duct. In types 2 and 3, ERCP images show both major and minor pancreatic ducts communicating with each other.

Our patient had type 1 total agenesis of dorsal pancreas, as MRCP was showing only part of major pancreatic duct in the ventral bud of pancreas. There was no evidence of any chronic pancreatitis.

The etiology of the abdominal pain of congenital abnormality of the pancreas is unknown and has been presumed to be due to pancreatitis or autonomic neuropathy similar to that in patients with diabetes mellitus.⁵

Diabetes was present in most of the reported cases. In some cases it substantially preceded the onset of abdominal discomfort and the diagnosis.⁶ Most patients with congenital short pancreas with normal situs had diabetes mellitus and were occasionally associated with pancreatitis.

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Multiple gastrointestinal and extragastrointestinal stromal tumors in a male infant — an extreme rarity

Introduction

GISTs are defined as submucosal mesenchymal neoplasms of the gastrointestinal tract, having spindle cell, epithelioid or pleomorphic morphology and characteristic positive immunostaining for CD117 antigen.¹ Until recently, they were regarded as smooth muscle neoplasms and categorized as leiomyosarcomas, leiomyomas or leiomyoblastomas.² In 1983, Mazur and Clark coined the term stromal tumors to describe the group of mesenchymal neoplasms that lacked ultrastructural and immunophenotypic features of smooth muscle differentiation.³

GISTs in pediatric age group are rare and constitute about 1% to 2% of all GIST cases and they mostly occur in females with a female to male ratio of 9:1.⁴ These are located most commonly in stomach (50–60% of lesions) and less commonly in small intestines (20–30%), large intestine (10%) and esophagus (5%).¹ There have been reports of GISTs occurring in extra gastrointestinal locations, primarily the mesentery, omentum and retroperitoneum, but these are rare. They are thought to originate from the interstitial cells of Cajal, the pacemaker cells of gastrointestinal tract like both of them express the receptor tyrosine kinase (KIT) which stain with

CD117 antigen.² Over expression of KIT due to activating mutations seems to be the trigger factor for neoplastic growth of GISTs.^{5,6}

So, it can be emphasized that multiple pediatric gastrointestinal stromal tumors occurring simultaneously at gastrointestinal (terminal ileum) as well as extragastrointestinal (mesentery and omentum) locations in a male infant is a rarity.

Case Report

A six months old male child with normal birth history and uneventful neonatal period was brought to the Accident and Emergency department with complaints of pain abdomen, vomiting, distention abdomen and non passage of stools for 4 days. There was no history of bleeding per rectum / malena. On examination, the child was febrile, tachypnoeic, had tachycardia (pulse rate-120/min) and normal blood pressure. On abdominal examination, the abdomen was distended and there was a single hard lump of about 9 × 6 cm present in the right iliac quadrant which was movable to some extent along two axes, perpendicular to each other. The child was investigated and an X ray abdomen showed multiple air fluid levels suggestive of intestinal obstruction (**Figure 1**). Ultrasonography revealed a soft tissue tumor in the right lower quadrant of the abdomen arising from the small intestine. CT scan of the abdomen was not done in view of urgency due to acute intestinal obstruction.

The child was taken up for surgery and on exploration, there was an extra-luminal hard tumor of size 8 × 6 × 5 cm on the anti-mesenteric border of the terminal ileum about 5 cm proximal to ileocaecal junction causing obstruction due to compression resulting in dilatation of whole of the small intestine proximal to the tumor (**Figure 2**). In addition, there were multiple small rounded hard tumors in mesentery (n=2) and greater omentum (n=3) varying in diameter from 0.5 cm to 1.5 cm. The liver and all other viscera appeared to be normal. About 7cm segment of terminal ileum, bearing the tumor, along with its mesentery and a margin of 2cm was resected and all the small tumors in omentum and mesentery were also excised (**Figure 3**) and end to end ileo-ileal anastomosis was done.

The patient died of septicemia on 4th post operative day. On histopathological examination, grossly the tumor was oval, homogenous, grey white, firm and on anti-mesenteric surface of ileum. Microsections from all the tumors (the large one as well as all the small ones) showed features of gastrointestinal stromal tumors comprising of spindle cells as well as epithelioid